

Linear atrophoderma of Moulin

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Key words: atrophoderma; linear hyperpigmentation; lines of Blaschko.

INTRODUCTION

Linear atrophoderma of Moulin (LAM) is a rare and distinct clinical entity characterized by acquired unilateral, hyperpigmented, and atrophic bandlike skin lesions following the lines of Blaschko. We report a rare and interesting case of LAM.

CASE REPORT

A 22-year-old Malay woman presented with an 11-year history of asymptomatic, unilateral brown patches affecting the right arm, right side of the trunk, and right leg. There were no prior skin lesions or inflammation. There was no significant medical or family history.

Physical examination found linear hyperpigmented atrophic patches on the right arm, right trunk extending down to the right buttock, right thigh, and right leg following Blaschko's lines involving both anterior and posterior aspects (Fig 1, A). The skin was slightly atrophic on palpation. No signs of induration or inflammation were noted (Fig 1, B).

Laboratory investigations including full blood count, erythrocyte sedimentation rate, renal profile, liver function test, antinuclear antibodies, double-stranded antinuclear DNA antibodies, anti-SCL-70 antibody, anti-SSA(Ro), anti-SSB(La), anti-Jo-1, and anti-RNP antibodies were all negative or within the normal range.

The first skin biopsy from a hyperpigmented patch over the right arm found mild upper dermal perivascular lymphocytic infiltrate. No dermal sclerosis was seen. Skin biopsy was repeated from normal skin (Fig 2, A) and from the hyperpigmented patch (Fig 2, B) on the right arm. Histopathologic examination of the affected skin found a mild upper dermal perivascular lymphocytic infiltration. When compared with the adjacent normal skin, the dermal thickness was reduced and the dermal collagen appeared more compact. The sweat glands,

Abbreviation used:

LAM: Linear atrophoderma of Moulin

pilosebaceous units, and appendages were not affected.

The diagnosis of LAM was made. Our patient was treated with betamethasone, 0.1% cream twice a day for 6 months, with no improvement. She then tried hydroquinone 4% cream for 2 years of which also failed to improve the hyperpigmentation.

DISCUSSION

Moulin et al¹ first described LAM in 1992, of which only 5 cases were observed over a 17-year period. Baumann et al² reported on a further patient with similar features and proposed the name *linear atrophoderma of Moulin*.²

The diagnostic criteria for LAM³ include

1. Onset during childhood or adolescence
2. Development of hyperpigmented, slightly atrophic, unilateral lesions following Blaschko lines on the trunk or limbs
3. Absence of preceding inflammation and absence of subsequent induration or scleroderma
4. Stable, nonprogressive clinical course without a pattern of remission
5. Histologic findings showing hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibers

Although different from the original report of Moulin et al,¹ the most common histologic finding is a perivascular lymphocytic inflammatory infiltrate in the superficial dermis combined with abnormal collagen fibers.³

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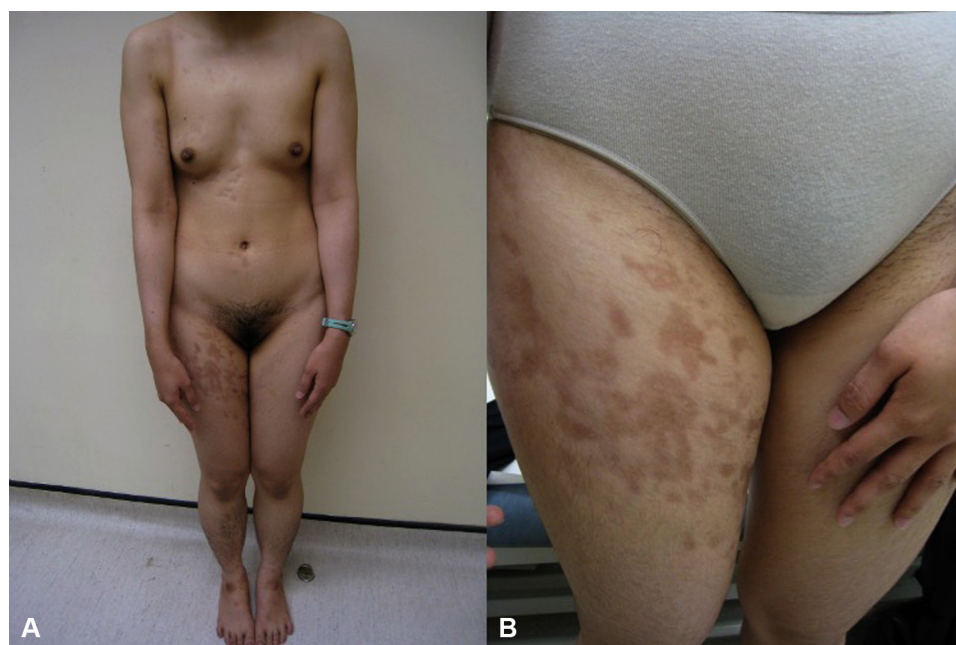


Fig 1. **A**, Linear hyperpigmented atrophic patches on the right upper inner arm, right side of the upper chest, right thigh and right leg following Blaschko's lines. **B**, Close-up view of the atrophic patches on right thigh.

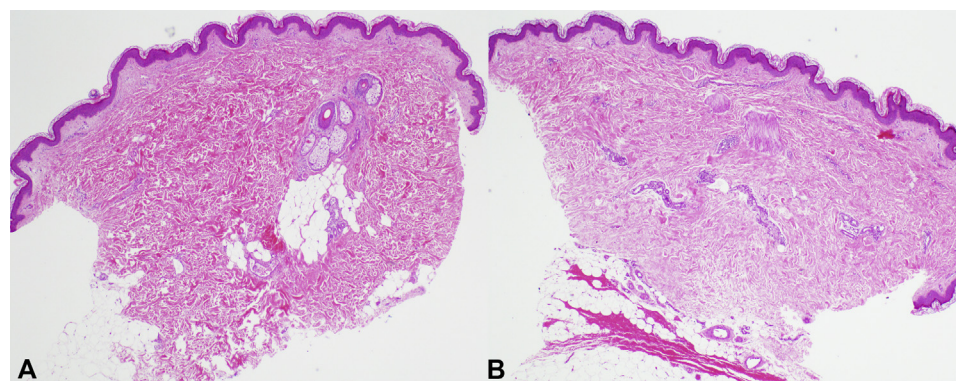


Fig 2. **A**, Histopathology of the normal skin. **B**, Histopathologic examination of the affected skin found reduced dermal thickness with more compact dermal collagen and mild upper dermal perivascular lymphocytic infiltration. (Hematoxylin-eosin stain; original magnification: $\times 40$.)

The etiology of LAM remains unknown. All reported cases were so far sporadic. Dermatoses that follow Blaschko's lines are thought to be caused by a somatic mutation that takes place early in embryogenesis, resulting in a genotypic and phenotypic mosaicism. Danarti et al⁴ postulated that LAM may reflect the action of an autosomal lethal gene surviving by mosaicism. A postzygotic mutational event may lead to loss of the corresponding wild-type allele at the atrophoderma locus, and this gives rise to a homozygous cell clone which becomes manifested along the lines of Blaschko later in life.

The differential diagnosis of LAM includes congenital dermatopathy following Blaschko lines such as linear and whorled nevoid hypermelanosis, incontinentia pigmenti, lichen striatus, and epidermal nevi. LAM should also be differentiated from atrophoderma of Pasini and Pierini, which also presents with similar configuration, atrophy, and hyperpigmentation but does not follow Blaschko lines. Atrophoderma of Pasini and Pierini is considered an abortive variant of morphea. In our patient, the initial clinical diagnosis was linear morphea. However, the clinical presentation,

absence of preceding inflammation, induration, or scleroderma and absence of dermal sclerosis on histopathologic examination, led to a diagnosis of LAM. It is important to differentiate LAM from linear morphea, as the prognosis is different. The prognosis of LAM is favorable, as there are no associated complications.

There is no effective treatment for LAM. Topical corticosteroids and heparin² were not helpful. High dose penicillin⁵ and potassium aminobenzoate⁶ were noted to be ineffective as well. A partial response to topical calcipotriol was reported.⁷ A case of LAM successfully treated with methotrexate 20 mg/wk for 6 months with an improvement of pigmentation and atrophy was reported.⁸ The authors suggested that LAM, atrophoderma of Pasini and Pierini, and linear scleroderma may be a spectrum of a common disease entity in view of the response to methotrexate, which is an effective therapeutic option for morphea. However, some cases of LAM do not respond to potent topical steroids and topical calcipotriol, which are the first-line treatment for morphea.

Since the original description, much debate has occurred regarding whether atrophoderma of Pasini and Pierini is a distinct entity or an abortive, nonindurated variant of morphea. Subsequent reports of the co-occurrence of morphea and occasionally lichen sclerosus with atrophoderma of Pasini and Pierini suggest a close relationship between atrophoderma of Pasini and Pierini and morphea. However, Yokoyama et al⁹ reported that skin glycosaminoglycans extracted from atrophoderma of Pasini and Pierini lesions are different from those in typical morphea lesions.

It remains controversial whether LAM is a distinct entity or on a spectrum of a common disease entity with atrophoderma of Pasini and Pierini and

linear scleroderma. Although some similarities exist between these 3 conditions, there are also differences in the age of onset, distribution, histology, origin, development, and prognosis of the lesions.

This case highlights the importance of recognizing the clinical presentation of this disease with atrophic hyperpigmented patches along Blaschko lines. When atrophoderma is suspected, skin biopsies should be taken from both normal skin and lesional skin, as atrophoderma often shows very subtle features.

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